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Balloon Dilation of Pulmonary Venous Pathway Obstruction After Mustard Repair for Transposition of the Great Arteries

STEPHEN G. COOPER, MB, FRACP, IAN D. SULLIVAN, MB, BMedSc, FRACP,
CATHERINE BULL, MB, MRCP, DCH, JAMES F.N. TAYLOR, MD, FRCP, FACC

London, England

Percutaneous balloon dilation was attempted in three patients with mid baffle pulmonary venous pathway obstruction after the Mustard operation for transposition of the great arteries. The procedure was unsuccessful in a 3 year old boy. Evidence for relief of obstruction in the other two patients (7 and 14 years old, respectively) consisted of angiographic demonstration of improved caliber at the site of the pulmonary venous pathway narrowing, improvement in the Doppler spectral signal at this site from an "obstructed" to a "normal" pattern as well as symptomatic improvement. Balloon dilation was performed twice in one of these patients. The Doppler and symptomatic improvement were sustained in both patients at short-term follow-up (5 and 6 months, respectively). This technique may offer effective relief of pulmonary venous pathway obstruction in some patients with this complication of the Mustard operation.

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The Mustard operation was, for many years, the preferred operation for physiologic repair of transposition of the great arteries. Complications of this operation include both systemic and pulmonary venous pathway obstruction (1). Reoperation is recommended in cases of pulmonary venous or inferior vena cava pathway obstruction and symptomatic superior vena cava pathway obstruction (2). Successful percutaneous balloon dilation of systemic venous pathway obstruction has been reported (3). We have employed balloon dilation in three cases to attempt relief of pulmonary venous pathway obstruction.

Methods

Patients. Since December 1987, pulmonary venous pathway obstruction has been identified in three children attending The Hospital for Sick Children. Each had undergone a Mustard repair for simple transposition of the great arteries. All three patients had presented with neonatal cyanosis and had undergone balloon atrial septostomy at that time.

Patient 1 underwent, at age 4 months, a Mustard operation in which native pericardium was used for the baffle and polytetrafluoroethylene (Gore-Tex) for right atrial enlargement. Superior vena cava obstruction at the site of the baffle, recognized at age 3 years, was repaired with the use of xenograft pericardium, with subsequent relief of facial edema. At age 7 years, the patient complained of tiredness, shortness of breath and nocturnal cough. Chest radiographs showed pulmonary venous congestion with moderate cardiomegaly.

In Patient 2, a Mustard repair was performed at another institution at age 4 days. He made an uncomplicated recovery from this operation and has been asymptomatic with no unexpected clinical findings. However, at age 3 years, chest radiographs showed pulmonary venous congestion.

Patient 3 underwent at age 6 months a Mustard procedure in which native pericardium was used to fashion the atrial baffle. The postoperative course was uncomplicated. At age 14 years, he reported having had increasing shortness of breath on exertion during the preceding year. Chest radiographs demonstrated mild cardiomegaly with pulmonary venous congestion.

Doppler echocardiography. In each patient, two-dimensional echocardiography suggested obstruction due to narrowing in the middle of the pulmonary venous pathway with dilation of the pulmonary veins. Doppler flow patterns at the site of pulmonary venous pathway obstruction were compared before and after balloon dilation with the use of an Ultramark 8 system. These studies were performed without

From the Cardiothoracic Unit, The Hospital for Sick Children, Great Ormond Street, London, WC1N 3JH, England.

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Address for reprints: I. D. Sullivan, MB, Cardiothoracic Unit, The Hospital for Sick Children, Great Ormond Street, London, WC1N 3JH, England.

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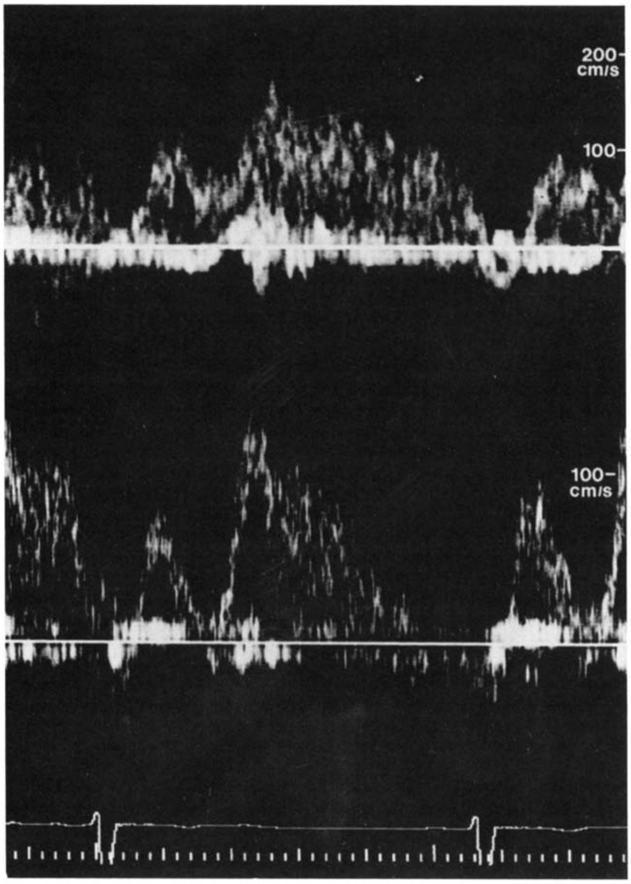


Figure 1. Patient 1: Pulsed Doppler signal at the site of pulmonary venous pathway obstruction. **Upper panel**, Before balloon dilation, maximal flow velocity is elevated and velocity remains high throughout diastole. **Lower panel**, After dilation, maximal flow velocity is diminished and flow is no longer continuous throughout diastole.

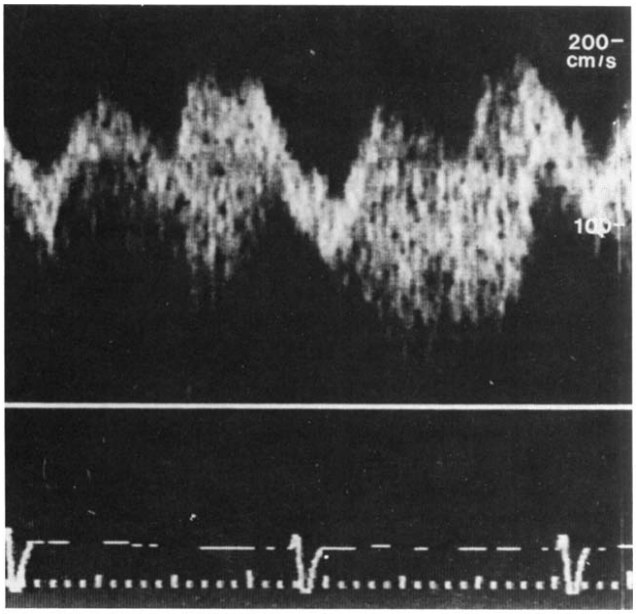


Figure 2. Patient 2: Pulsed Doppler signal at the site of pulmonary venous pathway obstruction showing continuous flow at high velocity throughout the cardiac cycle.

sedation during quiet respiration. Characteristic flow profiles with increased maximal flow velocity and spectral broadening, indicative of pulmonary venous pathway obstruction (4), were obtained in each patient before intervention (Fig. 1, top; Fig. 2). The maximal flow velocity and diastolic slope of the spectral signal were used to measure "pressure half-time" by analogy to flow at an atrioventricular valve (5). Maximal flow velocity occurred coincident with the expected timing of the y descent of the pulmonary venous atrial pressure waveform (6), as demonstrated previously (4). The mean value recorded over three representative cardiac cycles was documented.

Balloon catheter technique. Informed consent from the patient or his or her parents was obtained in each case. The procedures were performed under general anesthesia. Arterial and venous catheterization were performed percutaneously, and the diagnosis of pulmonary venous pathway obstruction was confirmed by hemodynamic and angiographic assessment. An end hole catheter was passed retrogradely by way of the tricuspid valve and positioned in the left upper or lower pulmonary vein. This catheter was then exchanged for a balloon catheter with use of an exchange guide wire (Fig. 3). The balloon size was chosen so that the inflated balloon area was equivalent to a circle with a diameter three to four times the dimension of the narrowest part of the pulmonary venous pathway measured by echocardiography (3,7). The balloon was inflated by hand with dilute contrast medium for 5 to 10 s. Balloon inflation was repeated four or five times in Patients 1 and 3; Patient 2 tolerated a single balloon inflation very poorly and it was not repeated.

Results

Immediate results. In Patients 1 and 3, the pressure difference between the pulmonary vein and the segment of the pulmonary venous pathway above the tricuspid valve was virtually abolished. This improvement was confirmed by Doppler assessment 1 day after the procedure (Table 1). There was also considerable improvement in the angiographic appearance at the stenotic site (Fig. 4A and B). The procedures were uncomplicated and both patients were discharged home the next day. In Patient 2, the narrowing was so severe that passage of even the deflated balloon resulted in severe bradycardia. A single balloon inflation was performed. Despite prompt withdrawal of the balloon catheter, restoration of cardiac output required external cardiac massage and administration of epinephrine. After the proce-

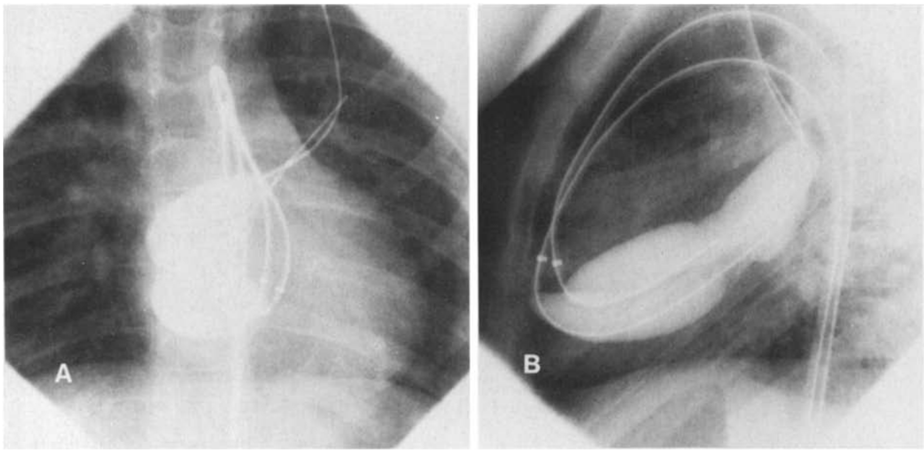


Figure 3. Patient 1: Anteroposterior (**A**) and lateral (**B**) angiographic frames showing two balloon catheters inflated at the site of pulmonary venous pathway obstruction. The tips of the exchange guide wires can be seen in the left upper pulmonary vein.

dure the patient's femoral pulse was difficult to feel but returned to normal with intravenous streptokinase. He was discharged home the next day.

Follow-up. *Patient 1* was reassessed 6 months later. A Doppler study suggested some residual pulmonary venous pathway narrowing (Table 1). Despite the lessening of the patient's symptoms it was believed that the balloon size used previously might have been inadequate, judging by the size of the balloons required for effective dilation of systemic venous pathway obstruction (3). The procedure was repeated with simultaneous use of two balloon catheters, one introduced through each femoral artery. This procedure was followed by angiographic (Fig. 4C), hemodynamic (Fig. 5) and echocardiographic evidence for further relief of obstruction (Table 1). Apart from the development of a groin hematoma, there were no complications. At 3 months of further follow-up, the pulmonary venous pathway Doppler spectral signal was similar to that obtained immediately after the second balloon dilation procedure, with a pressure half-time of 130 ms. At 6 months the reported improvements in effort tolerance and the abolition of nocturnal cough were sustained.

Patient 2 remained asymptomatic, but Doppler studies at 1 month follow-up demonstrated residual severe obstruction. It was believed that the obstruction was too severe for successful balloon dilation and the patient was referred for surgery.

In Patient 3 the Doppler signal at 3 month follow-up was similar to that obtained 1 day after the dilation procedure, with pressure a half-time of 127 ms. He continued to report improved exercise tolerance 5 months after the procedure.

There was Doppler evidence of mild tricuspid regurgita-

Table 1. Summary of Three Cases

	Patient 1: (first procedure)		Patient 1: (second procedure)		Patient 2		Patient 3	
Age (yr)	7.5		8.3		3		14	
Weight (kg)	26.2		26.4		13.5		50	
Min. PVP diameter (echo) (mm)	4		5		3		6	
Balloon (s)	18 mm × 5.5 cm		15 mm × 5.5 cm × 2		12 mm × 4 cm		15 mm × 5.5 cm × 2	
	Pre	Post	Pre	Post	Pre	Post	Pre	Post
$\overline{PV}$ (mm Hg)	32	26	14	18	29	24	27	32
$\overline{PVA}$ (mm Hg)	26	24	12	15	20	22	15	30
RVEDP (mm Hg)	11	14	7	10	8	11	10	24
Pressure half-time (ms)	280	170	165	120	300	320	339	125
$\dot{V}_{max}$ (m/s)	1.6	1.3	1.3	1.1	1.8	1.8	1.5	1.2

Min. = minimal; Post = after angioplasty; Pre = before angioplasty; $\overline{PV}$ = mean pulmonary vein pressure; $\overline{PVA}$ = mean pulmonary venous atrium pressure below mid baffle obstruction; PVP = pulmonary venous pathway; RVEDP = right ventricular end-diastolic pressure; $\dot{V}_{max}$ = maximal Doppler flow velocity at mid baffle obstruction.

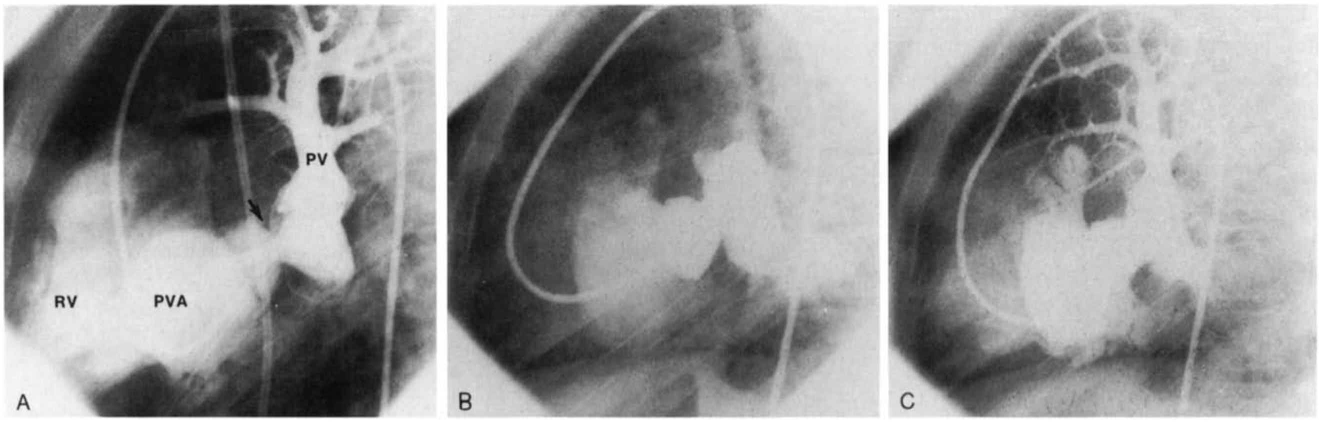


Figure 4. Patient 1: Lateral angiographic frames before dilation (**A**), after the first dilation procedure (**B**) and after the second dilation procedure (**C**). There is progressive improvement in the dimension of the localized obstruction (**arrow**) within the pulmonary venous pathway. PV = pulmonary vein; PVA = pulmonary venous atrium below the site of obstruction; RV = right ventricle.

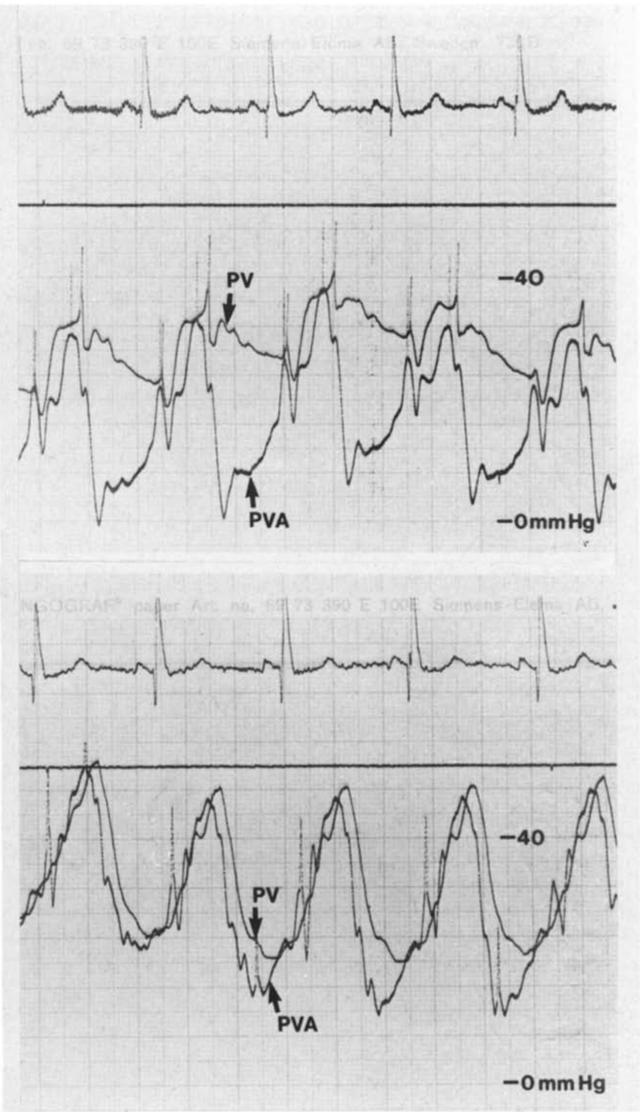
tion before the first balloon dilation procedure in Patient 1, and this was unchanged at serial evaluation. Patients 2 and 3 had no evidence of tricuspid regurgitation.

Discussion

The use of the Senning operation in preference to the Mustard operation as the atrial redirection procedure of choice has reduced the incidence of venous pathway obstruction (8), and the arterial switch procedure avoids potential venous pathway obstruction. Despite these changes in technique, there are still many survivors of the Mustard procedure at risk of this "late" complication. The Mustard operation is still being performed successfully in the current era (4). Consequently, pulmonary venous pathway obstruction will continue to be identified.

Comparison with surgery. Surgery has been the treatment of choice for pulmonary venous pathway obstruction complicating a Mustard repair. This approach has potential problems. Surgery may not abolish the obstruction and recurrent or residual narrowing may occur (2). Dysfunction of the systemic right ventricle is a well recognized complication of the Mustard operation. A second bypass procedure, and in some cases even a third procedure (see Patient 1), may well contribute to further deterioration in the function of the systemic right ventricle. Pooled data from six series (2) demonstrate that mortality at reoperation for pulmonary venous pathway obstruction is >20% even in experienced hands. These facts, together with the successful use of balloon dilation for systemic venous pathway obstruction (3), led us to attempt this technique for mid baffle obstruction of the pulmonary venous pathway.

Figure 5. Patient 3: Simultaneous pressure measurements above (PV) and below (PVA) the site of localized pulmonary venous pathway obstruction before (**upper panel**) and after (**lower panel**) balloon dilation. Abbreviations as in Figure 4.



Evaluation of results. In all three patients the pressure difference across the stenotic site was reduced immediately after balloon dilation. In Patient 2 this reduction was not indicative of relief of obstruction, as demonstrated by the Doppler findings before and the day after the procedure (Table 1); presumably, it reflected temporarily reduced cardiac output. In the other two patients there was almost complete abolition of the pressure difference between the pulmonary vein and that part of the pulmonary venous pathway above the tricuspid valve, and angiographic, Doppler and symptomatic changes suggesting lessening of the pulmonary venous pathway obstruction.

Conclusions. Our data suggest that this procedure may be effective in the short term without major complications. Some patients will not be suitable because of their small size or because of severity of the pulmonary venous pathway obstruction, but in our two older patients the short-term results have been encouraging. Our limited experience to date suggests that repeated balloon dilation may be effective if a suboptimal result is achieved in the first instance.

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